

A case of overlapping primary sclerosing cholangitis and autoimmune hepatitis presented as acute on chronic liver failure

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7-14% of patients suffering from primary sclerosing cholangitis (PSC) may present with clinical and serological features, indicating an overlap syndrome with autoimmune hepatitis (AIH). In an otherwise indolent course of a chronic liver disease, an exacerbation can result in acute liver failure (ALF). A 22-year-old man with a history of thrombotic events and verified thrombophilia (PAI-1 mutation) presented with jaundice (bilirubin=211,3 µmol/L) and abdominal discomfort. His synthetic liver function rapidly deteriorated (AST=3084 U/L, ALT=2928 U/L, ALP=154 U/L, GGT=63 U/L). Initial workup excluded viral causes and he was referred to the University Hospital Merkur with the diagnosis of ALF of unknown etiology. Whilst pursuing the underlying cause of the ALF and concomitant anaemia, the diagnosis of Crohn's disease was established via lower endoscopy. In the following days, the patient's state severely worsened, including the development of stage 3 hepatic encephalopathy. 7 days after admission, the patient was successfully transplanted. Histological assessment confirmed the diagnosis of overlapping AIH and PSC. The patient was uneventful for 2.5 years, when after the rise of liver enzymes (AST=296 U/L, ALT=682 U/L, GGT=93 U/L, ALP=165 U/L), a liver biopsy confirmed the relapse of AIH. Remission was attained with the administration of corticosteroids. In the 3-year follow-up, the patient was relapse-free without thromboembolic events. This complex case emphasizes the importance of prompt management of ALF and the difficulty of distinguishing acute on chronic liver failure from ALF. Overlapping liver diseases can be accompanied by various conditions, most commonly inflammatory bowel diseases, which makes the treatment particularly challenging.